ON HÆMATOMA OF THE DURA MATER ASSOCIATED WITH SCURVY IN CHILDREN.

BY G. A. SUTHERLAND, M.D. EDIN., M.R.C.P. LOND.

Physician to the North London Consumption Hospital; Registrar and Clinical Assistant, Paddington Green Children's Hospital.

THE manifestations of scurvy in children present some differences from those seen in the case of adults. The most important of these are the absence of sponginess of the gums, and the presence of extensive sub-periosteal hæmorrhages in connection with the long bones. The explanation of these variations is to be found, I believe, in the fact that rickets is almost always present, and leads to considerable alterations in the vascular supply of those parts in which scorbutic changes are most marked. In rachitic infants the growth of the teeth is at a standstill, due probably to a diminished blood supply, and sponginess of the gums may not appear during an attack of scurvy. As regards the bones, in rickets there is a greatly increased vascular supply, as shown by the marked periosteal activity, and the thickening around the epiphyses. If scurvy supervene, the hæmorrhagic tendency will naturally show itself in those places where the vascular supply is greatest, and consequently periosteal and epiphysial hæmorrhage is a prominent sign. For a similar reason the active changes present in the skull in rickets seem likely to lead to hæmorrhage in that region if scurvy is present. The occurrence of hæmorrhage under the pericranium has been noted by several observers. With the exception of a doubtful case of Möller's, quoted by Dr. Barlow, I have not met with any recorded case of infantile scurvy in which intracranial hæmorrhage was present. My own experience is limited to the following two cases which

were admitted into Paddington Green Children's Hospital. I have to express my indebtedness to Dr. Leslie Ogilvie for permission to publish these notes, and to Dr. Leonard Guthrie for his reports of the *post-mortem* examinations.

Case 1.—Female child, aged two years : anæmia ; fracture of humerus and femur ; periosteal effusion ; bruise-like extravasations ; death ; autopsy ; hæmorrhage into the brain and its membranes.

This patient was admitted to hospital on March 21. The child had been healthy when born, and was fed on breast milk entirely for four months. Since that time her food had consisted almost entirely of Nestlé's milk. There had been no snuffles or rash observed. Four months previously she had been put out to nurse. The nurse stated that in the beginning of February the child seemed to have great pain in the right thigh, which was swollen, and she took her to a hospital where disease of the hip joint was diagnosed. Two days before admission the mother got the child back, and noticed a swelling along the right thigh, and also along the left upper arm. There was also a bruise-like discolouration over the right side of the forehead.

The patient was an only child. The father and mother were healthy, and there had been no miscarriage.

On admission the child was found to be fairly well nourished, and had a pale waxy complexion. There were some dirty looking sores along the upper lip, and *alæ nasi*, and also on the buttocks. A large bruise-like mark was present on the right frontal eminence, greenish in colour. The signs of rickets were well-marked. The anterior fontanelle was open, tense and pulsating. The ribs were beaded, the epiphyses of the long bones were enlarged, and the bones of the forearm were curved. On the left upper arm a smooth, firm, tender swelling occupied rather more than the middle third. The swelling felt like periosteal thickening. The right thigh was bent at an angle a little above the centre, and the femur was fractured. The upper fragment was directed forwards and the lower backwards. The lower part of the shaft was much thickened in its whole extent. No crepitus was elicited between the fragments, but there was extreme tenderness over the whole thigh, and the child cried with a shrill, piercing scream when the limb was touched or moved. The temperature was 96° and the pulse 100 per minute, small and weak. There was nothing abnormal in the heart, lungs or abdomen.

On *March* 23 (two days after admission) signs of cerebral affection were present. The child was sick after food or medicine. Pulsation had ceased in the fontanelle, and the pulse was weaker. There was rigidity of the neck. The pupils were dilated, and the head and eyes were directed strongly to the left side. The left side of the face was drawn up. Both arms were rigidly flexed, and the hands were supinated; there was a good deal of tremulous movement in the arms at times. The child sank gradually, and died the same day.

Necropsy.—The body was fairly-well nourished. A subcutaneous hæmorrhage was present over the right frontal region, and another over the front of the right thigh. The skull was markedly rachitic. On opening the dura mater there was found a large amount of blood, dark and fluid, covering the vertex on both sides, from the middle of the frontal to the posterior part of the parietal regions. The surface of the brain was covered with a layer of blood clot, and the cortex was deeply stained. The blood also extended between the hemispheres. The structures at the base of the brain were covered by a thick, yellowish membrane, which was very tough, and contained no tubercles or nodules. It extended about half-way upon each side, and passed into the Sylvian fissures.

The brain contained two hæmorrhagic cavities under the cortex, situated in the posterior frontal region, one on each side. That on the right side was the size of a small Tangerine orange, and contained grumous, sanious fluid. The cavity on the left side was smaller and more irregular, and was partly filled with a small quantity of yellow, turbid fluid. Under the microscope, this fluid was seen to consist of blood-cells, mixed with a considerable number of fattily degenerated corpuscles. The lateral ventricles contained from two to three ounces of clear fluid. The extravasation under the dura mater seemed to be of some standing, from the considerable amount of organisation present in the membrane covering the base of the brain. The arteries at the base of the brain seemed thickened. On microscopic examination, the the basilar artery was found to be occupied by a thrombus which had undergone some degree of organisation.

The right femur was broken across almost transversely immediately above the centre. There was no attempt at union of the fragments, which were considerably separated. A large amount of soft material was lying round the shaft, under the periosteum, for the most part unorganised, but of a cartilaginous consistence in parts. A similar condition was present in the left humerus,

there being no appearance of union between the fragments, and a large quantity of soft material surrounded the broken ends of the shaft. The liver was tough, rather wrinkled and small. The other organs presented no abnormal appearances.

Case 2.—Female child, aged 14 months : rickets ; œdema of eyelids ; anæmia ; cachexia ; hæmorrhages in skin, scalp, and conjunctiva ; ulceration round teeth ; tenderness and œdema of extremities ; syncopal attacks ; death ; autopsy ; hæmatoma of dura mater.

This patient was seen for the first time on November 11, when she was ten months old. She had had a fit five days previously, which had left her with convergent strabismus. Abdominal pain seemed to be present. The nose and hands were blue, and the mother stated that the child was subject to cold extremities. She was wasted and seemed to have been neglected. The skull was large, and there was distinct bossing in the frontal and parietal regions. The anterior fontanelle was widely open, and the sutures had not closed. The heart was normal, and a good deal of bronchitis existed in the lungs. The abdomen was prominent, and both liver and spleen were enlarged. There was no history of hæmaturia. The patient was suffering from "snuffles," but there was no other evidence of syphilis, nor could any history of it be obtained. Her diet at the time consisted of cow's milk.

As regards the family history, both parents were healthy. There had been twelve children in the family, of whom only three survived, the others having died young, one from jaundice, and the rest from bronchitis. There had been no miscarriages.

The patient had always been subject to bronchitis. She never had fits with the exception of the one mentioned. In the end of the following January the child was seen again. Both upper eyelids were œdematous. She was suffering from another attack of bronchitis, and looked ill. She was admitted to hospital on February 10.

On admission, the patient was moderately well nourished, but markedly anæmic, with a yellow translucent skin. Over the right parietal bone, and part of the occipital, was a soft fluctuating swelling, considered hæmatomatous. The head was large and natiform, with distinct bosses. The anterior fontanelle was open, and the wrists were rather thickened. There were two subcutaneous hæmorrhages on the back about the size of a shilling piece, and another on the inner surface of the right arm. There was no œdema of the extremities. The right arm was kept rigid,

and the thumb was firmly adducted; there was also some tendency to adduction of the left thumb. The tongue was furred and dry, and there was some ulceration round the teeth. The liver extended one inch, and the spleen one-and-a-half inches, below the costal margin. The heart was normal, and there was a good deal of bronchitis. The patient was unable to swallow, and had to be fed with the stomach tube. The temperature was 100°, and the pulse 120, rather weak. The urine contained a trace of albumin, but no blood.

On February 12 she had a syncopal attack: the extremities became blue, the pulse could not be felt, and the pupils were contracted. She recovered under stimulation.

On February 15 she seemed to be improving. Her colour was not so yellow. She was still very apathetic, although not unconscious, and cried out whenever she was moved. As a rule she laid perfectly still, moaning at times as if in pain. The thighs were drawn up and adducted.

On March 7 an eruption of hæmorrhagic spots appeared on the abdomen and the back, for the most part small in size. This was accompanied by a rise in temperature, which had been normal for some days, to 101.4°, and by diarrhœa, the motions being green and offensive.

There was another syncopal attack on March 9. On the 13th there were numerous fresh petechial spots over the trunk. The hands and feet were swollen, and the lower limbs seemed very tender to touch. There was a marked increase in the diarrhœa, which had been more or less present during the previous week.

She died on March 26. The child had been lying in a state of torpor since the last note. At times there was a condition of general tonic spasm of the muscles, the eyes being turned to the right. The extremities were blue and œdematous, and the cardiac action was very feeble. The abdomen was extremely retracted. The temperature had been gradually failing, with occasional pyrexial attacks, and for two days before her death it was 95°. No treatment had any effect. The diet consisted of fresh cow's milk, milk puddings, lime juice, and brandy, and medicinally she had grey powder, mercurial inunction and cod liver oil.

Necropsy.—The body was extremely emaciated, with a complete absence of subcutaneous fat. A subconjunctival hæmorrhage was present in the inner and upper surface of the right eye. The teeth and gums were covered with sordes. On the

mucous membrane of the lower lip opposite the central incisors, was an ulcer, deeply excavated, with hard, raised edges, and a yellowish base.

The skin was wrinkled and parchment-like. The anterior surface of the body, from the nipple line to the umbilicus was densely spotted with purple, *ante-mortem* petechiæ, mostly the size of a pin's head. There were a few *ante-mortem* bruise-like marks about the extremities, of larger size than those on the trunk, notably one on the outer side of the right leg, and two symmetrically placed on the back of each forearm, the size of a threepenny-piece.

There was a considerable anasarca of both feet and ankles, and clear fluid escaped on puncturing these regions. A more solid œdema occupied the backs of both hands. There were no ulcers or excoriations about the arms or buttocks.

The skull was typically natiform. The parietal and frontal eminences were very protuberant, and the fontanelle was open and depressed. The epiphyses of the long bones were also somewhat enlarged. Incisions made over the tibiæ showed the bones to be normal. There were no periosteal thickenings, and no subperiosteal hæmorrhages. There was no epiphysitis anywhere in a more advanced condition than that mentioned above.

There was some collapse in both lungs, and pleuritic adhesions were present on the right side. The liver was small, mottled on cut section, and there seemed to be some increase of Glisson's capsule along the portal vessels. The spleen, kidneys and intestines appeared normal.

On dissecting off the scalp, a ragged red appearance was noted on the inner surface covering the right parietal bone. A large hæmatoma had been situated here three weeks before death. The parietal and frontal eminences, and also the squamous portions of the temporal bones, were much thickened, of a deep red colour, and soft enough to be indented easily by the finger-nail.

Beneath the dura mater there was in the situation of the subdural space, a dense, fleshy, purplish red, smooth and soft deposit, which completely covered the surface of the brain. It was generally loosely adherent to the under surface of the dura mater, and could be easily peeled off, except in the neighbourhood of the sinuses, where it was more firmly attached. The dura mater above it was white and glistening. The deposit varied from one-sixteenth to nearly three-quarters of an inch in thickness, the thickest parts being towards the base of the skull in the anterior and posterior fossæ. Beneath this layer, and easily

CASE 2.—Vertex of skull shewing false membranes *in situ*. The different layers, with cotton wool pledgets interposed, are seen on the left side of the skull anteriorly.

separable from it, was a second dense white membrane, and on the under surface of this was a third layer of soft, yellowish, coagulated lymph, somewhat closely adherent to it, and to the pia mater beneath. On removing this third covering, the brain was exposed, and presented the following unusual appearances. It was generally atrophied, and weighed fifteen ounces. The convolutions were ill-defined, and narrow, and the sulci were obscured, particularly the fissures of Sylvius and Rolando, by a layer of yellowish solid lymph, encircling the contracted and anæmic blood-vessels, and causing the pia mater to bulge externally.

The convolutions generally were of a pale greyish colour. In striking contrast to these were certain limited convolutions, which were of an entirely different aspect. The latter were of a dirty orange yellow colour, considerably depressed below the general surface of the convolutions, feeling so hard and dry to the touch that they were thought at first to be the seat of old caseating gummata. However, on section, it seemed that only the pia mater and the extreme outer surface of the grey matter were thus affected. The pia mater was thickened and gritty, and perhaps the yellow appearance was due to a fatty degeneration of old inflammatory deposits.

The various cranial nerves were white and glistening, and to all appearance normal. There was no thrombosis of the sinuses, and no disease of the petrous, or any other bone. On section, the brain was found to be tough, but otherwise normal.

On opening the spinal canal, there was a deposit of soft material like red currant jelly beneath the dura mater, and covering the cord, particularly well-marked in the posterior dorsal region. It seemed justifiable to regard this as an effusion of blood-stained lymph, and as perhaps resembling the early stage of the much more advanced effusion in the cranial subdural space.

Microscopic Examination.—The false membranes around the brain presented the appearance of blood clot in different stages of organization. In some parts they consisted of fibrinous tissue containing unchanged blood-cells, in others organization was more advanced, and strands of connective tissue were found, forming spaces which contained groups of altered blood-cells.

In the liver there was a slight increase in the amount of connective tissue surrounding the portal vessels, but the hepatic cells were unaltered.

Remarks on Case 2.--In this case we have a combination of diseases. The signs of rickets and scurvy are, I think, unmistakable, and there is also a certain amount of evidence in favour of syphilis. The distinct bossing of the skull is by some regarded as distinctive of syphilis, while by others, among whom is to be included Dr. Barlow, it is accounted a manifestation of rickets. The question has not been definitely settled, and is not of special importance in this case, for while the possibility of syphilis being present may be admitted, the whole series of symptoms, and the pathological changes found at the *post-mortem* were not such as are present in syphilitic disease, but were quite in accordance with a diagnosis of rickets and scurvy. Further, mercurial treatment had been employed without benefit, both while she was attending as an out-patient, and after her admission to hospital.

Remarks on Cases 1 and 2.--In both of these a leading feature was the condition known as pachymeningitis interna hæmorrhagica, or hæmatoma of the dura mater. This was extremely well marked in the latter case, and was present to a slighter extent in the former.

The best account of this affection is given by Huguenin in Ziemssen's "Cyclopædia of the Practice of Medicine" (vol. xii.) He says: "Pachymeningitis occurs in scurvy; here, however, we have no personal observations to offer. Many cases exist where syphilis is supposed to be the original disease; the connection is not clear; certainly no one will be disposed to refer the affection to Heubner's disease of the cerebral vessels."

In Dr. Immerman's article on scurvy in Ziemssen's Cyclopædia (vol. xvii.), which is extremely full and comprehensive, the occurrence of intracranial hæmorrhage is regarded as very exceptional amongst adults. He says: "As very rare local affections of scurvy are to be considered the isolated cases of intermeningeal hæmorrhage, which usually run their course under the form of rapidly-fatal apoplexy, but also in some cases lead to death with the appearance of gradually augmenting compression of the brain, headache,

dulness, and finally stupor ; it is still doubtful whether or not hæmorrhages occur in the substance of the brain." He does not record any case with *post-mortem* examination.

Huguenin refers to Virchow's theory that pachymeningitis is a hæmorrhagic inflammation of the dura mater, but is of opinion that inflammation is never present, and that the whole process consists of extravasation of blood, at first from the dura mater, and subsequently from already existing false membranes.¹ As regards the pia mater, he has frequently seen " the changes attributed to recurring fluxionary hyperæmia (formation of small fibrous callosities, or at times a thick, dense, leathery membrane, in connection with atrophy of the brain)." He has also observed " a yellow colouration of the arachnoid, of the subarachnoid fluid, and even of the surface of the brain, penetrating to a greater or less depth." Amongst all the cases that Huguenin was able to collect, he found that 2·7 per cent. occurred in children under twelve months, and 2·7 per cent. in children from one to ten years of age.

These appearances recorded by Huguenin are very similar to the ones I have described in the two cases under consideration. About the nature of the meningeal changes there can be no doubt ; the only question is as to the primary disease, for hæmatoma of the dura mater is always a secondary affection. In the history of the cases I have adduced the evidence on which I base the diagnosis of scurvy. In hæmatoma of the dura mater we find :

(1) A tendency to recurrent attacks of hæmorrhage without local inflammation.

(2) That coagulation and organization of the extravasated blood occur, and

(3) That the effects are entirely those of mechanical pressure.

These conditions are also characteristic of the subperiosteal and other hæmorrhages of scurvy.

In Case 1 there was no evidence during life as to the

¹ Dr. Wigglesworth also supports the non-inflammatory view. *Vide* BRAIN. Parts lix. and lx., 1892. " Remarks on the Pathology of so-called Pachymeningitis Interna Hæmorrhagica."

period of onset of the meningeal affection, but the organized membrane at the base of the brain pointed to an attack some time previously. In Case 2 the occurrence of a fit at the age of ten months, accompanied by strabismus, points to the first extravasation of blood from the dura mater. Accepting this as our guide, we find that the patient lived four months after the commencement of the pachymeningitis.